

8B

## Ventilation for motor neurone disease (MND)


### Information for people with or affected by motor neurone disease

If your breathing grows weaker with motor neurone disease (MND), it is important to think about whether you wish to use assisted ventilation, where a machine helps support your breathing.

This information sheet explores what to think about when deciding whether to use this support. It includes:

- 1: **Why might I need my breathing to be assisted?**
- 2: **What do I need to think about with non-invasive ventilation (NIV)?**
- 3: **What do I need to think about with tracheostomy ventilation (trache ventilation)?**
- 4: **How do I look after my ventilation equipment?**
- 5: **What do I need to know about stopping assisted ventilation?**
- 6: **How do I find out more?**

**This sheet contains sensitive information from the beginning about stopping ventilation and planning ahead for future care. Reading about this may feel difficult, but may help you make timely decisions about treatment and care in the future. Read when you feel ready to do so.**

 This symbol is used to highlight **our other publications**. To find out how to access these, see *Further information* at the end of this sheet.

 This symbol is used to highlight **quotes** from other people with or affected by MND.



This information sheet has been endorsed by the Association of Chartered Physiotherapists in Respiratory Care (ACPRC).

**This information has been evidenced, user tested and reviewed by experts.**

## What do the words mean?


When you discuss assisted ventilation with health and social care professionals, some words can sound similar, or different words may mean the same thing. This can be confusing. The following definitions may help:

<b>assisted ventilation</b>	Where a machine supports your breathing.
<b>BiPAP</b>	BiPAP stands for Bilevel Positive Airway Pressure, and is a type of non-invasive ventilation. You may hear some people use this term instead of NIV.
<b>dependent</b>	Being dependent on ventilation means you can no longer breathe effectively without it. If you need to use a ventilation machine for more than 12 hours in every 24, it usually means you are becoming more dependent on it. This can happen with either NIV or trache ventilation. You may also hear this called being 'reliant'.
<b>mouthpiece ventilation (MPV)</b>	A form of NIV that lets you 'top up' your breathing by putting a straw-like nozzle into your mouth when you want to pump extra air into your lungs. Ask your respiratory team about this if you think it may be useful.
<b>non-invasive ventilation (NIV)</b>	A type of assisted ventilation, where the machine boosts the flow of normal air into your lungs, usually through a mask held on by straps around your head.
<b>tracheostomy</b>	The surgical procedure to make an opening in the front of your neck, where a tube can be inserted into your windpipe. This can then be used for ventilation.
<b>tracheostomy ventilation (trache ventilation)</b>	A type of assisted ventilation, where the machine blows normal air into your lungs through a tracheostomy tube. You may hear people call this 'invasive ventilation'. For ease of reading, we will call this 'trache ventilation' for the rest of this sheet (where 'trache' is pronounced as 'tracky').
<b>respiratory care or management</b>	Breathing support and therapies.
<b>respiratory team</b>	The care team involved in supporting you with breathing problems.
<b>secretions</b>	Mucus and saliva.

# 1: Why might I need my breathing to be assisted?


With MND, your lungs can usually still process air. However, muscles weaken, including those that pull air in and out of the lungs. Over time these muscles stop working, but a machine can help support your breathing to relieve a number of breathing symptoms and prolong survival.

This is called assisted ventilation. For ease of reading, we will shorten 'assisted ventilation' to 'ventilation' for the rest of this sheet. See sections 2 and 3 of this sheet for information on the types of ventilation used to support breathing with MND.

 For detailed information on how MND affects breathing, see: Information sheet 8A – *Support for breathing problems*

In most cases, ventilation for MND uses normal air to help you breathe. Extra oxygen is not usually recommended with MND, as it can upset the balance in your body between oxygen and carbon dioxide. However, oxygen may be needed with MND if you have a chest infection or another condition that affects your lungs. It is also sometimes used during end of life care to provide comfort and ease.

## When do I need to think about ventilation?

 *"Forewarned is forearmed to what may lie ahead. I always feel it better to know what's ahead and ways of dealing with it. The unknown is always more frightening."*

Deciding whether or not to use ventilation can be a difficult choice. It can help you to make informed decisions by having open and honest conversations with your respiratory team about:

- your options for ventilation
- the benefits of each option and things to think about
- what is likely to happen over time with each type of ventilation
- what care is like after ventilation is introduced
- any other questions you may have.

This decision will also depend on:

- your needs
- your views, preferences and wishes for your future care
- assessment to see if the treatment is suitable for you
- the best time for ventilation to be introduced.

Find out as much as you can about the options for ventilation as soon as you feel ready. It is very important to have discussions with all those involved in your care before you develop any breathing related symptoms, or at the first sign of any changes to your breathing.

This will give you time to:

- think about your decisions, without a sense of urgency
- understand what your choices mean, now and in the future
- ensure those close to you and your health and social care team know your wishes and preferences, to help you access the treatments you want and avoid unwanted or unplanned treatments or interventions
- ensure everyone who supports you is prepared for the changes ahead, whether or not you decide to use ventilation.



*“My own experience has shown that people are not always prepared early enough. Or symptoms have not been spotted early enough. In my husband’s case, his breathing problems were too far advanced for him to use non-invasive ventilation effectively.”*

These conversations may raise questions about work and finances for you and your family. Your health and social care team can support you with these conversations.

Early planning with your respiratory team about how you wish to use ventilation can help you find the best way to support your breathing, which may include part-time use at first. This can also help you and those supporting you organise your care, which may help you stay at home to be cared for. Emergency decisions about ventilation can make this more difficult.

## **What are the early signs I may need ventilation?**




*“When I visit the MND clinic every three months I have a vital capacity test and a cough test. I am sure my care team would step in and give me advice and recommendations as and when they observed my lung capacity declining to a worrying level.”*

The following can be signs of weakened breathing muscles, which may mean you need ventilation or other support for breathing:

- feeling breathless, even when resting
- shallow breathing, even when lying flat
- difficulty clearing mucus or phlegm
- a weak cough
- repeated or long lasting chest infections

- a weak sniff
- disturbed sleep
- feeling tired when you wake up or during the day
- nightmares or hallucinations
- morning headaches
- poor concentration, poor memory or feeling confused
- voice getting weaker
- having less appetite for food, which can cause you to lose weight.

 For more information on breathing tests and symptoms with MND, see: Information sheet 8A – *Support for breathing problems*


## How much support is needed with ventilation?

Either type of ventilation can prolong survival compared to not using ventilation to support breathing. Historically, trache has always been thought to prolong survival longer than NIV, but research is ongoing into ventilation survival times with MND. As findings become clear, we will update this information sheet.


Although ventilation can prolong survival, it will not stop the breathing muscles becoming weaker or slow the wider progress of the disease. This means that your breathing can be supported, but your other symptoms will get worse over time. You will need more support as the disease progresses.


If you have not yet been referred to a speech and language therapist (SLT), ask for a referral, as weakened breathing muscles can affect the way you speak and communicate.

You may reach a point when you can no longer move or communicate without help. The time this takes varies from person to person. Access to computers through eye movement and adapted switches can provide communication support.

 *"I use a computer controlled by my eye movements to surf the web, read and write emails, and even use spreadsheets."*

Simple communication aids can also be used if needed, for example, where changes to thinking or eye problems make it difficult to use high-tech communication aids.

 *"I wish we'd known earlier of eye-movement aids which might have been helpful to him. Unfortunately, he's too ill now to be able to make use of them."*

 For information on speech, communication and how to record your voice to use a computerised version of it in the future, see information sheets:  
7C – *Speech and communication*  
7D – *Voice banking*

MND can affect the way you eat, drink and swallow. Using ventilation can also make eating and drinking difficult. It is important to think about whether you wish to use a feeding tube to help you get enough nutrients into your diet. Now is a good time to open conversations with your family, carers and health and social care team about tube feeding, as this can become more difficult to introduce when the breathing muscles become weaker.

You will need support from a speech and language therapist (SLT) and a dietitian, to help manage swallowing difficulties caused by MND and ensure you are getting enough nutrients.



For more detail, see:  
Information sheet 7A – *Swallowing difficulties*  
Information sheet 7B – *Tube feeding*  
*Eating and drinking with motor neurone disease*

Because MND can affect the way you swallow, food, drink and saliva may be at risk of 'going down the wrong way'. With a weak cough, this may be difficult to clear.

When food, liquid or saliva gets into your lungs, it is called 'aspiration'. If this happens to you, get assessed by a speech and language therapist as soon as possible to look at the way you swallow and provide support.

Aspiration can cause chest infections. You may notice:

- a runny nose or increase in mucus
- increased breathlessness
- a sore throat or a cough
- symptoms of fever or just feeling unwell.

Contact your GP as soon as possible if you think you may have a chest infection, as early antibiotic therapy can help. You can also get help to cough, including:

- support from your health and social care team
- techniques you can learn yourself
- a machine that can help you cough.

Ask your doctor about the yearly flu and one-off pneumonia vaccines, as they can reduce the risk of getting these infections. These are usually available from your GP surgery. Your main carer and close family may also be able to get these vaccinations.

Because you can prolong survival with MND by using ventilation, your breathing muscles will weaken over time, and you are likely to become dependent on it. This can happen with either type of ventilation, but the time this takes varies depending on your symptoms and the type of ventilation you use.

Care needs become complex if you are dependent. Over time, you are likely to need 24 hour support from an experienced multidisciplinary team and all those involved in your care.

If you wish to be cared for at home, you are likely to need a team of people to be there 24 hours a day to provide support. Usually these will be paid care workers, but sometimes family members will want or need to take part in this, too.



*"I sleep on a daybed beside him so that I am on hand quickly when he needs his secretions suctioning in the middle of the night. I can't leave him alone in the room at any time."*

If MND or your ventilation equipment makes speaking or communicating difficult, ventilation systems come with alarms that can let people know if you need help. However, somebody needs to be there at all times to respond. Ask your health and social care team for guidance.

Family members and carers are likely to need reassurance, training and additional support for either type of ventilation, especially in the later stages of the disease.



*"We have a nurse who comes once a week to be with him so I can go and do a shop or go to any appointments. The rest of the time I am pretty much confined to the house. I'm not even able to hang the washing out as I wouldn't be able to hear any of his emergency alarms indicating a problem."*

The need for 24 hour support means that arranging a package of care can be difficult. Depending on how healthcare support works in your area, you may only be able to get the right level of support if you move into a nursing home. This varies across regions. Discuss this with your respiratory team, who can support you in finding out what the options are in your area, and planning how you will use ventilation to support your breathing.

Everyone has a different experience with MND. Some people adjust to increasing disability and wish to extend life for as long as possible. Ventilation can help prolong survival and a combination of care support and assistive aids can help to improve quality of life. However, if you wish to make a different choice based on your preferences or beliefs, that is your right and should be respected. Discussion of all the support and options available may help you come to a decision that is right for you.

## **What happens if I decide not to use ventilation?**



*"It's important to know that ventilation can increase life expectancy with MND as, in our case, my husband did not want to extend his life."*

The decision to use, or not use, ventilation is your choice. You may decide you don't want to use ventilation, or find you cannot adjust to its use. If so, your respiratory team, physiotherapist or palliative care team can advise on other ways to help ease symptoms.

These may include:

- changing your posture and positioning
- breathing exercises
- help to cough
- finding ways to relax
- medication to ease breathlessness and anxiety.



For detailed information about other breathing therapies, see:  
Information sheet 8A – *Support for breathing problems*

Although this support can help ease symptoms, they will get worse over time, and breathing therapies cannot slow the progress of the disease. Eventually, this is likely to lead to death. However, your health and social care team can support you with medication to ease anxiety.

## **MND Just in Case Kit**

One of the ways that may help you feel reassured with breathing difficulties is by having an *MND Just in Case Kit* nearby.

The *Just in Case Kit* is a box you can keep in your home, which contains medication for use in emergency situations. Having the kit nearby can be reassuring should you become breathless or experience coughing or choking.

We supply the kit box free of charge to your GP, at their request. Your GP then prescribes suitable medicine for you to keep in the box at home. This ensures medication is on hand for a doctor or community nurse, if they are called out of hours.

Your GP may need to include a signed permission for other professionals to use the medication.

The box also contains written guidance for professionals. If your GP feels it is appropriate, they may also prescribe medication for your carer to give to you while you are waiting for professional medical help to arrive.

Your GP can contact our helpline, MND Connect, for advice about the kit. See *Further information* in section 6: *How do I find out more?* for contact details.



## 2: What do I need to think about with non-invasive ventilation (NIV)?

Non-invasive ventilation (NIV) is commonly used to support breathing problems with MND. It supports your breathing by boosting the flow of normal air into your lungs, usually through a mask held on by straps around your head.

The mask is attached by a tube to a machine called a ventilator. The machine is powered either by electricity or battery, or both. Your breathing triggers the machine, so that when it senses you taking a breath, it provides a supporting boost of air to help you breathe effectively.

A range of masks are available, but usually cover your nose, or your nose and mouth. If masks are difficult to use, nasal pillows may be available, which deliver air through two small tubes that sit inside the nostrils and are held on by straps around your head (you may hear some people call these 'nasal prongs').

The settings, mask and other parts of the ventilator will be adjusted to suit you. This is usually carried out at hospital as an outpatient, but may require a short stay. However, some of these services may be able to visit you at home if you find it difficult to travel.

It may be possible for current models of ventilation equipment to be adjusted by your respiratory team remotely, even if you live far away from the hospital. Ask your health and social care team to find out the options in your area.





*"My NHS NIV pump sends data to my ventilation team for analysis. Based on the findings, they can adjust any of the settings – including how humid the air is!"*

NIV is a recommended treatment for breathing difficulties in the National Institute for Health and Care Excellence (NICE) guideline on MND (NG42). NICE guidelines set out recommendations for health and social care professionals on treatment and care for people living with certain conditions.



For more information on the NICE guideline, see:  
Information sheet 1A – *NICE guideline on motor neurone disease*

and our pocket sized booklet on opening conversations with professionals:  
*What you should expect from your care*

Possible benefits of using NIV	Things to think about with NIV
<p data-bbox="185 241 268 331"></p> <p data-bbox="280 241 836 398"><i>“Wearing my [NIV] ventilator 24 hours a day allows me to lead what I call a ‘normal’ life and doesn’t stop me from doing what I want to do.”</i></p> <ul data-bbox="197 448 826 1411" style="list-style-type: none"> <li>• it can ease symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches (this can make you feel better during the day and have more energy to do the things you want to do)</li> <li>• it can increase survival time</li> <li>• it is portable and can be used in many places, including at home and on the move, if powered by battery</li> <li>• surgery is not needed for NIV</li> <li>• although you are likely to need more support over time, NIV care is generally quite easy to manage in the earlier stages of MND</li> <li>• a range of available masks means you can try what suits you best</li> <li>• using NIV can add volume to your voice if this has become weak.</li> </ul> <p data-bbox="185 1460 268 1541"></p> <p data-bbox="280 1460 820 1608">For information on flying while using ventilation equipment, see: <i>Information sheet 8D – Air travel and ventilation for motor neurone disease</i></p>	<ul data-bbox="877 241 1484 1630" style="list-style-type: none"> <li>• it can cause discomfort from air flow leaking around the mask and irritating the eyes</li> <li>• pressure of the mask on the skin can cause sores</li> <li>• wearing a mask can cause claustrophobia, which in some cases may lead to anxiety or panic</li> <li>• it can cause a blocked nose, a runny nose or dry mouth</li> <li>• you may need support with eating or drinking while using the equipment, as the flow of air may cause an increased risk of food or drink ‘going down the wrong way’ (known as aspiration)</li> <li>• it can make you swallow more air than usual, which can make your stomach feel full and uncomfortable, meaning it may take more effort to breathe</li> <li>• it can take a while to get used to the feel of the equipment and flow of air</li> <li>• care needs can be complex while using NIV in the later stages of the disease</li> <li>• the flow of air can disturb the rhythm of natural speech</li> <li>• although NIV machines are generally quiet, the noise can take some getting used to and may keep you awake at night at first, and anyone you share a room with.</li> </ul>

## What aftercare is needed with NIV?

Your respiratory team and wider health and social care professionals can assist if you experience any direct discomfort with the equipment.

Aftercare with NIV can be different for everyone, but may include:

- ensuring the mask is comfortable, eg by adjusting the position of the mask on your face and the tightness of the straps, or using different masks in rotation (dressings and skincare products advised by your health and social care team may also help)
- help to cough, to clear mucus and saliva (you may be able to learn techniques to do this yourself, or have support from your respiratory team or specialised equipment such as a cough machine)
- using eye lubricants in case of air leaking from the mask into your eyes (adjusting the fit of the mask may also help)
- adding a humidifier to the system, to add moisture to the air flow (this may help if you experience a dry mouth, runny nose or blocked nose)
- planning with your respiratory team and a speech and language therapist how to manage eating and drinking with NIV
- drinking more fluids, which can help with a dry mouth, thick or sticky saliva and constipation (some people find drinking pineapple juice helps with thick saliva)
- getting support from a speech and language therapist to help with communication difficulties, and alternative ways of communicating
- asking your respiratory team if your NIV equipment can be adjusted to reduce the risk of trapped air
- moving around, or being assisted to move, so that trapped air is helped to pass through your system (some people find drinking peppermint water also helps)
- getting support from your health and social care team to help with any feelings of anxiety or panic (see also *MND Just in Case Kit* in section 1).



For more details, see:

Information sheet 7A – *Swallowing difficulties*

Information sheet 7C – *Speech and communication support*

**The following section contains information about how the disease will progress. Reading about this may feel difficult, but may help you make informed decisions about ventilation. Read when you feel ready to do so.**

## **What happens over time with NIV?**

At first, you may only need to use NIV at night, to help improve your breathing and quality of sleep. As the disease progresses, you may also need to use it during the day to help relieve your symptoms.



*"I started just using NIV for an hour each day and slowly increased my use, so I found it easy when I needed to use it all night."*

Over time, you may become dependent on NIV. This is where you need to use it constantly and will be unable to breathe on your own without it. If this is the case, you should have a back-up machine that can be powered by battery (see section 4: *How do I look after my ventilation equipment?* for information on what to do in case of a power cut). You are likely to need increased care support, as needs can become complex. This might include overnight care.

The time it takes for someone to become dependent on NIV varies from person to person. For some people this can take months or years, for others it may happen more quickly.

As your breathing muscles weaken, your breathing will trigger the machine less often. However, the ventilator can deliver air through the mask if your breaths are not strong enough to trigger it for a set amount of time. When the machine is providing all your breaths and you no longer trigger it, it is called being 'fully ventilated'.

Because you can prolong survival with NIV, your other symptoms will continue to progress. With a weak cough, you may be at risk of secretions getting into your lungs, which is known as aspiration. This can lead to a chest infection called aspiration pneumonia, which can be life threatening if left untreated. Help with coughing can help reduce the risk of aspiration, and antibiotics can help treat infections, but there may come a time when these can no longer help.

### 3: What do I need to think about with trache ventilation?

With MND, trache ventilation supports your breathing using a tube inserted into your windpipe through an opening made in the front of your neck. This tube is connected to a ventilator machine, powered by electricity or a battery, or both.

The surgical procedure to make the opening in your neck is called a tracheostomy. The tube is usually inserted under general anaesthetic (meaning you will be asleep for the procedure). As well as supporting your breathing, the tube may also be used to suction airway secretions (see later heading, *What aftercare is needed for trache ventilation?*).

Trache ventilation is not always offered to people with MND in the UK. If you want to discuss whether trache ventilation is the right choice for you, ask your respiratory team about the options and support available in your area.

Your respiratory team is most likely to suggest trache ventilation:

- if you find NIV uncomfortable, or because your MND is too far advanced for NIV to be effective
- if the mouth and throat muscles are weak and there are a lot of secretions, as trache ventilation can be used to help manage this
- if more extensive suctioning is needed during a serious chest infection.






It is also sometimes used in an emergency to take control of your breathing (although, as it can be difficult to stop using trache ventilation with MND, you may want to have control over this decision, see later heading *Emergency situations*).

Following the tracheostomy, you will need a stay in hospital while a package of care is arranged for when you leave. The respiratory team will support you in hospital during this time. Arranging a care package can sometimes take months to organise, as finding or training a team of people (usually through a care agency) to cover care for a tracheostomy can be difficult. However, this varies across regions. Discuss this with your respiratory team, who can advise on the situation in your area.




*“Despite general caution about invasive ventilation, I decided to proceed with a tracheostomy after deliberations with my family and extensive reading of research papers.”*

It is important to have conversations about trache ventilation with your family, carers and health and social care team before you make any decisions.

Possible benefits of using tracheostomy ventilation	Things to think about with tracheostomy ventilation
<p data-bbox="151 212 742 459">  <i>“Despite some inconveniences and discomfort (more from coping with being bedridden rather than the tracheostomy itself), I have enjoyed life, and never regretted the decision, nor have my family.”</i> </p> <ul data-bbox="167 481 742 1657" style="list-style-type: none"> <li>• it can improve symptoms caused by weakened breathing muscles, such as shortness of breath, disturbed sleep, daytime sleepiness or fatigue and early morning headaches (this can make you feel better during the day and have more energy)</li> <li>• it can increase survival time</li> <li>• it can be used where NIV cannot successfully support your breathing</li> <li>• depending on the type of trache used, it can help to protect the airway from aspiration and reduce the risk of lung infections, which can be life threatening (discuss types of trache tube with your respiratory team)</li> <li>• it can be used to help with control of secretions</li> <li>• a mask is not used, so mask related issues, such as claustrophobia, can be avoided</li> <li>• it can move around with you, including while using a wheelchair, if powered by battery.</li> </ul> <p data-bbox="151 1713 742 1836">      For information on flying with ventilation, see: Information sheet 8D – <i>Air travel and ventilation</i> </p> <p data-bbox="151 1870 742 2128">  <i>“I was a very active and busy man. With invasive ventilation, I’ve had to adapt to a very different way of life, including being far more attentive to family and friends, and more reflective. I’ve also become an expert in films!”</i> </p>	<p data-bbox="766 212 1508 324">  <i>“In our case, speech, eating and drinking became impossible when my husband had a tracheostomy.”</i> </p> <ul data-bbox="782 347 1508 1892" style="list-style-type: none"> <li>• like any surgical procedure, a tracheostomy carries some risks associated with infection and aftercare. These will be explained to you before the procedure, and in most cases can be easily managed</li> <li>• having a trache tube in the neck carries some risk of discomfort and long term damage to the windpipe, although this is not common</li> <li>• you may need a long stay in hospital after the operation, while your care team is organised</li> <li>• in some cases, care may only be provided in a nursing home (ask your respiratory team how healthcare support works in your area)</li> <li>• it is important to include family and friends involved in your care when thinking about having a tracheostomy, as it is highly likely they will be involved in your care</li> <li>• you will need the tube changing regularly, as guided by your respiratory team, which some people find uncomfortable (this is usually done every month, but may be less often, depending on your needs)</li> <li>• depending on the type of trache used, it may affect your ability to speak, eat and drink if these have not already been affected by MND. You may need support to communicate in other ways, and use a feeding tube to safely get enough nutrients into your diet</li> <li>• it can significantly increase secretions, which will need regular clearance from the airway (some people find this uncomfortable as this is done while you are off ventilation)</li> <li>• it can take a while to get used to the feel of the equipment.</li> </ul> <p data-bbox="766 1937 1508 2072">      For more information on alternative ways of feeding, see: Information sheet 7B – <i>Tube feeding</i> </p> <p data-bbox="774 2128 821 2184">14</p>

**The following section contains information about how the disease will progress. Reading about this may feel difficult, but may help you make informed decisions about ventilation. Read when you feel ready to do so.**

## **What happens over time with trache ventilation?**

 *“Together, seven years on, we have seen our children grow into young adults, get married and we have become grandparents to two beautiful babies. So there have been some amazing highs while he has been on trache ventilation, but there have also been so many days in between them which have been really hard.”*

If MND has caused your breathing muscles to weaken, you are likely to become dependent on trache ventilation as soon as it is introduced. This means that if you cannot get used to the way the machine feels, you are unlikely to be able to return to breathing independently without this support. However it is your right to stop using ventilation if you wish. See section 5: *What do I need to know about stopping assisted ventilation?* for more information.

As your breathing muscles weaken, your breathing will trigger the machine less often. However the ventilator can deliver air if your breaths are not strong enough to trigger it for a set amount of time. When the machine is providing all your breaths and you no longer trigger it, it is called being ‘fully ventilated’.

It is essential to make sure you have a ventilator that can run on battery power, and that the battery is fully charged. You will also need a spare machine in case the one you are using breaks down (see section 4: *How do I look after my ventilation equipment?* for what to do in case of a power cut).

Because you can prolong survival with tracheostomy ventilation, your other symptoms will continue to progress. You may be at risk of repeated infections, which can be life threatening.

For ways to record your wishes and preferences for your care and treatment, see: *How do I plan ahead?* in section 5: *What do I need to know about stopping assisted ventilation?*

## **Emergency situations**

In emergency situations, healthcare professionals may introduce trache ventilation without your prior consent.

For example, if a person is resuscitated in an emergency, trache ventilation is sometimes used during recovery. However, with MND it can be difficult to stop using this type of ventilation once introduced, as the breathing muscles may have weakened in the meantime. You may then have to decide whether to continue with trache ventilation or have it withdrawn.



*“He was rushed into hospital as he was having breathing difficulties caused by a severe chest infection. He had a tracheostomy, as it was hoped that he would be able to support his own breathing, but this was not the case and he has been on a ventilator ever since.”*

If trache ventilation is something you definitely do not want introduced in any circumstances, you need to make this clear to all those involved in your care. You can do this by recording your wishes to guide people if you become unable to make or communicate decisions about ventilation for any reason in the future. In some cases, this guidance may be legally binding. See *How do I plan ahead for my future care?* in section 5: *What do I need to know about stopping assisted ventilation?*

## **What aftercare is needed with trache ventilation?**

Ongoing aftercare with trache ventilation is essential for your safety and wellbeing.

Aftercare for trache ventilation can be different for everyone, depending on your needs and symptoms, but may include:

- continuous warming and moistening (humidifying) of the air as it blows into the tube, which can be delivered by a machine attached to your ventilator
- regular clearance and suctioning of secretions from the airway (this may be uncomfortable in the early days after trache ventilation is fitted, and you may need time to adjust to the feel of it)
- help to cough, to clear secretions (your family and carers may be able to learn techniques to do this, or you may need support from your respiratory team or specialised equipment, such as a cough machine)
- using a nebuliser with the ventilation system to get a fine mist of sterile saline solution (salt water) into your lungs, which may help to clear mucus
- regular maintenance of the tube and daily care to prevent blockages, infections or skin breakdown around the tube site
- support from a speech and language therapist to help manage speech and communication difficulties and get advice on alternative ways of feeding. Getting support from a dietitian is also important, to ensure you are getting enough nourishment from your diet.

Your respiratory team will support you and provide guidance on aftercare with trache ventilation, as suited to your needs.



For more information, see information sheets:

7A – *Swallowing difficulties*

7B – *Tube feeding*


7C – *Speech and communication difficulties*




## 4: How do I look after my ventilation equipment?

Your use of the ventilator will be regularly reviewed in case your needs change. The respiratory team can provide ongoing support, including advice about maintaining and cleaning the equipment, and out-of-hours urgent support, if needed.

### What if there is a power cut?

 *"If you use a suction machine, a battery-operated machine is a must in case of power cuts!"*

A spare battery and back-up ventilator is essential if there is a power cut in your area. Keep batteries charged for any ventilation devices and other battery powered support, such as suction pumps.

 *"We were on the energy supplier's critical care list for urgent response in times of power cuts."*

Contact your energy provider about what support you may be able to get by signing up as a priority user to the Priority Services Register. As a priority user, you should be told about any planned power cuts in your area. If there is a long power cut planned and you are dependent on your ventilator, ask the service if they can provide you with a back up generator to use during this time, or if you will need to arrange for one yourself.

However, if a power cut is likely to last several days and you are dependent on ventilation, you may need to stay at another address until the power is reconnected. This is also recommended if you only use ventilation part-time.

If you have speech and communication difficulties and your life is in danger, you or your carer can text the emergency services instead of calling.

To do this, you need to register your mobile phone online with the Next Generation Text Service at:

[www.ngts.org.uk/how-to-use-ngt/contact-999-using-ngt.html](http://www.ngts.org.uk/how-to-use-ngt/contact-999-using-ngt.html)

### Will the equipment need servicing?

Yes. Whoever supplied you with the equipment will provide ongoing servicing.

## What can be done at home to help maintain the equipment?

Using either type of ventilation at home means you may need to keep a lot of equipment close to you at all times, for example:

- suction machine
- spare ventilator
- spare charged batteries
- cough machine
- humidifier
- replacement tubes, disposables, etc (for trache ventilation).

Regular maintenance of the equipment can help keep it in good condition.

You, your carers and family members will be trained in maintaining your equipment when it is fitted in hospital. Ask your respiratory team or the equipment provider to explain if there is anything you do not understand or if you have any concerns.

**Non-invasive ventilation (NIV):** Regular maintenance may include:

- cleaning the mask
- replacing filters
- checking the straps or cushioned areas on the mask are not worn, as these can cause leaks.

**Tracheostomy ventilation:** Regular maintenance may include:

- replacing the whole tube (also known as the cannula), usually once a month, which can be a minor procedure but does carry some risk. Some people find this uncomfortable or worrying because the tube is changed while you are off ventilation (paid care workers are usually trained in changing the whole tube, but your respiratory or ear, nose and throat team can also support you with this)
- cleaning or replacing the inner tube (inner cannula), which your family and carers can be trained in doing
- daily or more frequent care to prevent blockages, infections and skin breakdown around the tubing.

Whoever supplies the equipment will provide contact details for help with any technical difficulty. This will include an out-of-hours contact. Keep contact details in a place that is easy to access, including details for your respiratory team.

**The following section contains information about stopping ventilation and end of life. Read when you feel ready to do so.**

## 5: What do I need to know about stopping assisted ventilation?

### Can I stop using ventilation?

Yes. It is your legal right to ask for ventilation to be stopped at any time. This is often called 'withdrawal'. You may wish to stop using ventilation if you feel it is no longer helping or has become a burden to you.


This is not the same as asking someone to help you end your life, which is known as assisted suicide.

If you use ventilation only some of the time, you may choose not to put the machine back on after a gap in use. You are likely to need support to manage your symptoms, so planning how you will withdraw ventilation with your health and social care team is very important.

If you can no longer breathe on your own without using ventilation, death is likely to follow in a fairly short amount of time after the ventilator is withdrawn. You will be supported through this by your care team following careful planning. However, your decision to stop must be made with the clear understanding that **it will cause a significant risk to your life**.

Discuss this with your respiratory team or palliative care professionals, who can:

- explain how medication can help you feel calm and relieve distress when ventilation is withdrawn
- answer any questions you may have about stopping ventilation, including how to manage symptoms and how your family will be supported.

 For more detailed information on stopping assisted ventilation, see: Information sheet 8C – *Withdrawal of ventilation with MND*

### How do I plan ahead?

Using ventilation may raise questions and fears about the way MND will progress. This is an important time to open conversations with your family and health and social care team about your wishes and preferences for your future care.

You may find it helpful to discuss:

- what will happen in the later stages of MND, as knowing the facts can help reduce fear and help you plan ahead more effectively
- how open you are to trying new technologies that may help you continue to communicate with others when other forms of communication become difficult
- what the advantages and disadvantages are for choosing to have interventions that are likely to extend your life, according to your wishes and preferences (such as assisted ventilation or a feeding tube)
- your choices for end of life care
- your options for withdrawal of ventilation
- how to record your wishes about future care and planning advance decisions.

The thought of having these conversations may feel upsetting. However, it is important to discuss your future care as early as you feel able to. Complex conversations can become more difficult, as MND can cause:

- difficulty with speech and communication
- fatigue or extreme tiredness
- changes to thinking and behaviour, which are usually mild, but can be more severe. This can affect up to half of people living with MND, although this may increase in the late stages of the disease.

Having and recording these discussions is called advance care planning. You can use certain documents to record your wishes about your care and treatment, for example withdrawing ventilation.

These can be used to guide those involved in your care if you become unable to make or communicate specific decisions about this in the future. This helps professionals, and your family and carers, understand how you want to be supported. It also ensures health and social care professionals are guided as to how you want future treatments, interventions and care to be arranged. This includes stopping or not introducing treatments you do not want. In some cases, this guidance may be legally binding.

Your wishes may change as your symptoms progress. You can review and change your advance care planning documents at any time.



For more information, see our planning ahead resources:

- *End of life: a guide for people with motor neurone disease*
- *Understanding my needs*
- Information sheet 14A – *Advance Decision to Refuse Treatment (ADRT) and advance care planning*

Our MND Connect helpline can also provide practical guidance on future planning. See *Further information* in section 6: *How can I find out more?* for contact details.

## What else do I need to think about?

You may decide you do not want the ventilator withdrawn. This means it will continue to support your breathing. However, with either type of ventilation, you may still be at risk if you have repeated chest infections, which can be life threatening.

The team supporting you will make every effort to help you make and communicate decisions about your future care. However, they may need to make decisions on your behalf if you become unable to do this or to communicate.

You can guide their decisions in advance, by:

- discussing your preferences for future care with them
- recording your decisions and preferences in an advance care planning document, such as an Advance Decision to Refuse Treatment (ADRT)
- giving Lasting Power of Attorney (LPA) to a trusted friend or family member of your choice.

With any decision about future care or treatment, your healthcare team will consider the benefit and the harm of the treatment, and will consult with those who know what your preferences might be, such as members of your family.

If continuing with ventilation is not in your best interests, any decision about this will not be taken lightly. However, if a point is reached when ventilation is harming you or is not in line with your previous wishes, it is likely to be withdrawn. You will be given medication to help you to be comfortable and avoid distressing symptoms. In most cases end of life with ventilation can be made peaceful with the right care and treatment.



For more information on ADRT and LPA, see:  
Information sheet 14A – *Advance Decision to Refuse Treatment and advance care planning*

## 6: How do I find out more?

### Useful organisations

We do not necessarily endorse any of the following organisations, but have included them to help you begin your search for further information.

Contact details are correct at the time of print, but may change between revisions. If you need help to find an organisation or have any questions, contact our MND Connect helpline (see *Further information* at the end of this sheet for details).

#### EmergencySMS (Next Generation Text Service)

If you have speech and communication problems, you can text the emergency services. You will need to register your phone with the Next Generation Text Service using the following web address.

Website: [www.ngts.org.uk/how-to-use-ngt/contact-999-using-ngt.html](http://www.ngts.org.uk/how-to-use-ngt/contact-999-using-ngt.html)

#### GOV.UK

Online government advice benefits and welfare, including support for disabilities, for people living in England and Wales.

Website: [www.gov.uk](http://www.gov.uk)

#### Health and Care Professions Council (HCPC)

A regulatory body who keep a register of health professionals who meet their standards of practice.

Address: Park House, 184 Kennington Park Road, London SE11 4BU

Telephone: 0300 500 6184

Website: [www.hpc-uk.org](http://www.hpc-uk.org)

#### Health in Wales

Online help about NHS services and health boards in Wales (see also NHS Direct Wales).

Email: through the website contact page

Website: [www.wales.nhs.uk](http://www.wales.nhs.uk)

#### MND Scotland

Care, information and research funding for people affected by MND in Scotland.

Address: 2nd Floor, City View, 6 Eagle Street, Glasgow G4 9XA

Telephone: 0141 332 3903

Email: [info@mndscotland.org.uk](mailto:info@mndscotland.org.uk)

Website: [www.mndscotland.org.uk](http://www.mndscotland.org.uk)

### **The National Institute for Health and Care Excellence (NICE)**

An independent organisation providing national guidelines and advice to improve health and social care, including NG42 on the assessment and management of MND. The guideline includes a section on NIV.

Address: 10 Spring Gardens, London SW1A 2BU  
Telephone: 0300 323 0140  
Email: nice@nice.org.uk  
Website: **www.nice.org.uk**

### **NHS UK**

The main online reference for the NHS, including information on continuing healthcare.

Website: **www.nhs.uk**

### **NHS 111**

The NHS helpline if you need urgent, but not life-threatening medical help or advice. Available 24 hours a day, 365 days a year.

Telephone: 111 (England and some areas in Wales)

### **NHS Direct Wales**

Health advice and information service for Wales. Available 24 hours a day, 365 days a year.

Telephone: 0845 4647 or 111 (if available in your area)  
Website: **www.nhsdirect.wales.nhs.uk**

### **Health and social care Northern Ireland (NHS Northern Ireland)**

Online information on health and social care services in Northern Ireland.

Website: **http://online.hscni.net/**

### **NI Direct**

Online government advice on benefits and welfare, including support for disabilities, for people living in Northern Ireland.

Email: through the website contact page  
Website: **www.nidirect.gov.uk**

## **Reference list**

References used to support this information are available on request from:  
email: infofeedback@mndassociation.org

Or write to:

Information feedback, MND Association, Francis Crick House, 6 Summerhouse Rd,  
Moulton Park Industrial Estate, Moulton Park, Northampton NN3 6BJ

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## **Further information**

We provide the following information sheets related to this information that may be helpful:

1A – *NICE guideline on motor neurone disease*

3D – *Hospice and palliative care*

7A – *Swallowing difficulties*

7B – *Tube feeding*

7C – *Speech and communication support*

8A – *Support for breathing problems*

8C – *Withdrawal of ventilation with MND*

8D – *Air travel and ventilation for motor neurone disease*

10A-G – *Our financial support and social care range of information sheets*

14A – *Advance Decision to Refuse Treatment (ADRT) and advance care planning*

We also provide the following guides and tools:

*What you should expect from your care* – our pocket sized booklet using the NICE guideline on MND to open conversations with your health and social care team

*Living with motor neurone disease* – our main guide about MND and how to manage its impact



*Caring and MND: support for you* – comprehensive information for unpaid or family carers, who support someone living with MND

*Caring and MND: quick guide* – the summary version of our information for carers

*End of life: a guide for people with motor neurone disease* – our comprehensive guide to making decisions about future care and late stage MND, including advance care planning and advance decisions

*Understanding my needs* – a write-on tool to help you record basic notes about your needs and how you would like to be cared for, if admitted to hospital or a hospice.

You can download most of our publications from our website at:

**[www.mndassociation.org/publications](http://www.mndassociation.org/publications)** or order in print from the MND Connect team, who can provide additional information and support:

### **MND Connect**

Telephone: 0808 808 6262

Email: [mndconnect@mndassociation.org](mailto:mndconnect@mndassociation.org)

MND Association, Francis Crick House, 6 Summerhouse Rd,  
Moulton Park Industrial Estate, Moulton Park,  
Northampton, NN3 6BJ

### **MND Association website and online forum**

Website: **[www.mndassociation.org](http://www.mndassociation.org)**

Online forum: **<https://forum.mndassociation.org>** or through the website

## **We welcome your views**

We'd love to know what you think we're doing well and where we can improve our information for people with or affected by MND, or Kennedy's disease. Your anonymous comments may also be used to help raise awareness and influence within our resources, campaigns and applications for funding.

To feedback on any of our information sheets, access our online form at:  
**[www.smartsurvey.co.uk/s/infosheets\\_1-25](http://www.smartsurvey.co.uk/s/infosheets_1-25)**

You can request a paper version of the form or provide direct feedback by email:  
[infofeedback@mndassociation.org](mailto:infofeedback@mndassociation.org)

Or write to:

Information feedback, MND Association, Francis Crick House, 6 Summerhouse Rd,  
Moulton Park Industrial Estate, Moulton Park, Northampton NN3 6BJ

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